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IMAGE

Platypnoea-orthodeoxia syndrome due to a patent foramen oval with marked lipomatous hypertrophy

Syndrome de platypnée-orthodéoxie en rapport avec un foramen ovale perméable et importante hypertrophie lipomateuse du septum inter-auriculaire

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MOTS CLÉS

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An 80-year-old man was admitted with severe refractory hypoxaemia following right lung radiotherapy for pulmonary carcinoma. The hypoxia was enhanced by the upright position and was associated with cyanosis. Computed tomography and lung scintigraphy did not identify a pulmonary cause explaining the degree of hypoxaemia. Transthoracic echocardiography (TTE) showed mild aortic root dilatation (48 mm) and a highly mobile atrial septum. Contrast-enhanced TTE showed massive and spontaneous right-to-left shunting through a large patent foramen oval (PFO). Transoesophageal echocardiography during PFO closure identified a markedly thickened septum secundum due to lipomatous hypertrophy and a very thin and floppy septum primum (Fig. 1). Right-sided pressures were within the normal range. A 20-mm sizing balloon (NMT Medical, Boston, MA) engaged inside the tunnel showed an initial waist due to the septum secundum, but with further advancement no additional waist was identified suggesting a large PFO up to 20 mm (Fig. 2). A 35-mm Amplatzer® Cribiform occluder was implanted with excellent shunt resolution and immediate and sustained improvement in gaseous exchange (Fig. 3).

The Amplatzer Cribiform occluder is usually indicated for multiperforated inter-atrial septa, but has proven efficacy in the PFO setting. It can be considered in cases such as this as it covers both septa well, permits overstretching due to the nitinol construction, and allows stability testing before delivery. The unusual anatomy more closely resembles a true septal defect and consequently percutaneous closure is challenging

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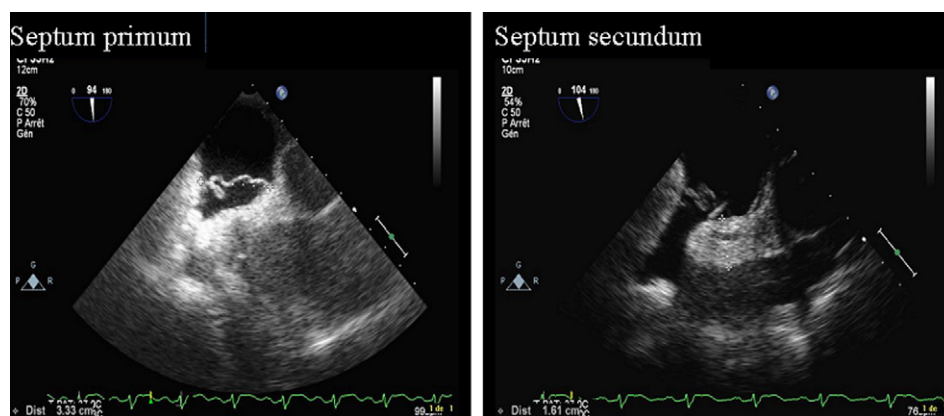


Figure 1. Atrial septal anatomy. Right panel: thick septum secundum (16 mm). Left panel: highly mobile thin floppy septum primum.

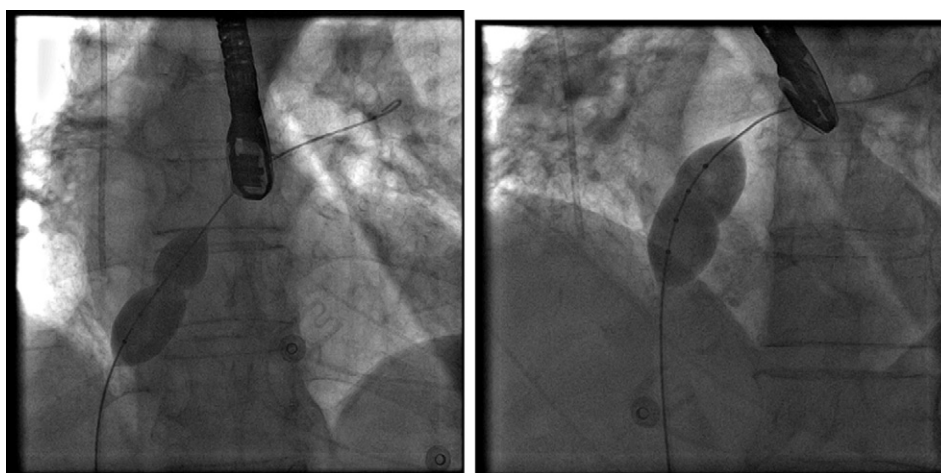


Figure 2. Patent foramen oval sizing, using a 20-mm NMT balloon. Right panel: no imprint when the balloon is advanced inside the PFO. Left panel: imprint of the septum secundum.

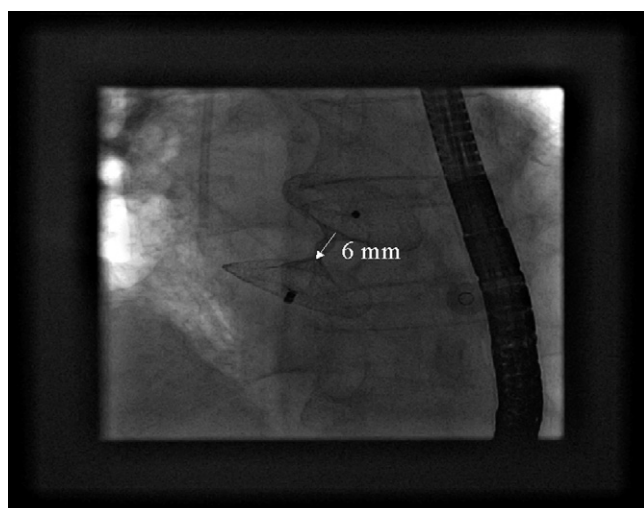


Figure 3. Amplatzer® Cribriform occluder device. Fluoroscopic overview of the implanted device overstretched since the distance between the two discs is 6 mm (usual distance is 3 mm). The diameter remained unchanged (35 mm).

and the selection of occluder closure device is critical. Our case nicely illustrates the effectiveness of this device with an unusual anatomy, and for this patient, it provided immediate symptom relief, allowing the patient to be discharged from hospital without the need for an oxygen supply.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.